CORONARY UNROOFING DOES NOT FITS ALL ANOMALOUS AORTIC ORIGIN OF CORONARY ARTERIES.

Luigi Arcieri[1], Massimo Colaneri[1], Francesco Bianco[1], Ettore Merlino[1], Gaetano Santoro[1], Raffaele Silvano[1], Alessandra Baldinelli[1], and Marco Pozzi[1]

[1]Azienda Ospedaliero Universitaria Ospedali Riuniti di Ancona Umberto I G M Lancisi G Salesi

February 22, 2024

Abstract

Introduction Anomalous aortic origin of coronary artery (AAOCA) is the second leading cause of sudden cardiac death in children and young adults. Intramural-interarterial course is the most frequent anatomic variation and coronary unroofing is widest adopted for surgical management. Symptoms recurrence is described regardless of the technique used. This study aims to describe how an anatomic patient-centered approach aimed to restore a normal coronary artery take-off is associated with symptoms resolution. **Methods** From 2008 to 2021, 25 patients were operated on for an AAOCA at a median age of 20 years. Nineteen patients had a right AAOCA and six had left AAOCA. Intramural course was present in 18 patients. Seventy-six percent were symptomatic. No episodes of aborted sudden cardiac death before surgery was described in the population. Surgical technique used were coronary unroofing in 18 patients, coronary neo-ostioplasty in 3, coronary re-implantation in 3 and main pulmonary artery re-location in one. **Results** No hospital mortality or re-operation was observed in our experience as well as major complications related to surgery. Mean hospital length of stay was 8.5 days. None of patients reported symptoms recurrence at follow-up. Young athletes returned to play competitive sport. Postoperative computed-tomography scan evaluation showed a general improvement of the take-off angle. **Conclusions** AAOCA requires a patient anatomic-based surgical management. There is not a single surgical technique that can fits all anatomic subtype of AAOCA. Surgical techniques may be selected on the base of the preoperative images and intraoperative findings. In our experience this policy is associated with no symptoms recurrence.

Introduction

Anomalous aortic origin of coronary artery (AAOCA) is the second leading cause of sudden cardiac death in children and young adults. Intramural-interarterial course is the most frequent anatomic variation and coronary unroofing is widest adopted for surgical management. Symptoms recurrence is described regardless of the technique used. This study aims to describe how an anatomic patient-centered approach aimed to restore a normal coronary artery take-off is associated with symptoms resolution.

Methods

From 2008 to 2021, 25 patients were operated on for an AAOCA at a median age of 20 years. Nineteen patients had a right AAOCA and six had left AAOCA. Intramural course was present in 18 patients. Seventy-six percent were symptomatic. No episodes of aborted sudden cardiac death before surgery was described in the population. Surgical technique used were coronary unroofing in 18 patients, coronary neo-ostioplasty in 3, coronary re-implantation in 3 and main pulmonary artery re-location in one.

Results

No hospital mortality or re-operation was observed in our experience as well as major complications related to surgery. Mean hospital length of stay was 8.5 days. None of patients reported symptoms recurrence at follow-up. Young athletes returned to play competitive sport. Postoperative computed-tomography scan evaluation showed a general improvement of the take-off angle.

Conclusions

AAOCA requires a patient anatomic-based surgical management. There is not a single surgical technique that can fits all anatomic subtype of AAOCA. Surgical techniques may be selected on the base of the preoperative images and intraoperative findings. In our experience this policy is associated with no symptoms recurrence.

Introduction

Anomalous aortic origin of coronary arteries (AAOCA) is a non-infrequent congenital, anatomic heart anomaly whose prevalence in the population is dramatically heightened in the last years [1-4].

The association with sudden cardiac death in athletes has improved the medical awareness of this malformation leading to an increase in cardiologic investigations, especially before competitive sport practice [5].

Several studies have attempted to quantify this value, with estimates ranging between 0.1 and 1% in both the adult and pediatric population. Anomalous aortic origin of right coronary artery (AAORCA) is from three to six times more frequent than anomalous aortic origin of left coronary artery (AAOLCA), that is, on the other hand more commonly associated with sudden cardiac death [6,7].

Surgical management of AAOCA continues to be a matter of debate especially in asymptomatic AAORCA [8-10]. Several surgical techniques have been developed in the last years with excellent results in terms of mortality and freedom from re-operations. Although the unroofing is the most used technique, neo-ostioplasty and coronary reimplantation are effective and have showed comparable outcomes [11-13]. Regardless of the technique used, is it the overall shape and the normalization of the take-off angle the mainstay of surgical management [13]. Also, unfortunately, there were some reports that have showed symptoms recurrence after surgery [14,15].

The aim of the present study is to describe our surgical management in AAOCA describing how an anatomic-based patient-specific approach is associated with complete symptoms resolution at mid-term follow-up.

METHODS

Study population and data collection

The study was approved by institutional review board of Ospedali Riuniti of Ancona. Patient consent for personal clinical data use was obtained. Patients enrolled for surgery for AAOCA between 2008 and 2021 at Pediatric Cardiac Surgery department of Ospedali Riuniti Ancona were included in the study. A retrospective review of data collected in an institutional database was made. Data recorded included demographics, instrumental, intra-operative surgical data, postoperative hemodynamic and clinical data. Follow-up was made after one, three and six months postoperatively and then yearly. Postoperative follow-up included clinical examination, echocardiography, exercise stress test and computed-tomography scan (at least after six months postoperatively).

Preoperative evaluation and indication to surgery

A standardized echocardiographic protocol has been developed in these years in our institution for the diagnosis of AAOCA [16]. Trans-thoracic echocardiography (TTE) and computed tomography scan were performed in all patients. Echocardiographic studies were performed using a commercially available Philips iE33 ultrasound systems (Philips Medical Systems, Amsterdam, The Netherlands), using a 2.5 or 3.5 MHz transducer, as appropriate. Images were acquired at a frame rate of 70–80 frames/s, applying a standard two-dimensional greyscale. Images of three consecutive heart cycles were collected from the parasternal short axis (PSAX) and long-axis (PLAX), and the 4/5-chambers apical views. Coronary computed tomography

examinations were subsequently obtained using a 384 (192Å˜2)-slices third-generation scanner (SOMATOM Force CT; Siemens Healthineers, Forchheim, Germany). The echocardiographic and CT images were evaluated and compared by dedicated team including radiologists with cardiac expertise and pediatric cardiologists with training in cardiac radiology. The images (Figure 1) were discussed with the team of surgeons before the planning of the operation and were re-discussed after the intra-operative observation in order to plan future cases.

The take-off angle of the anomalous coronary was calculated on CT axial view images, at the intersection of two lines of which one passed at the base of the coronary artery and the other in the first 5 mm of the vessel (Figure 2).

Indications to surgery were the presence of symptoms suggestive of ischemia (chest pain episodes especially during efforts, syncope or ventricular arrhythmia), the presence of an AAOLCA, the presence of an AAORCA with anatomic features considered at high risk (inter-arterial and long intramural course, i.e. more than 5 mm) regardless to symptoms especially in young athletes that rejected sport restriction.

Surgical technique

Surgery was made by anterior median sternotomy and cardiopulmonary bypass. Myocardial protection was obtained by a single initial infusion in the aortic root of the Bretschneider histidine-tryptophan-ketoglutarate (HTK) solution, commercially knows as Custodiol (Custodiol HTK, Köhler Chemie GmbH, Bensheim, Germany).

Surgical techniques employed were based on anatomic preoperative evaluation and intraoperative findings. The purpose of surgical management was the restoration of the best geometric take-off of the coronary from the aorta. In all cases a careful detection of the external course of the anomalous artery was made before aortic cross clamp in order to define the spatial relationship of the coronaries with the surrounding structures and to localize the point where the anomalous artery left perpendicularly the aortic wall (defined the real take-off).

Surgical unroofing was attempted in cases of AAOCA with intramural course. The aorta was opened and the length and direction of the intramural segment within the aortic wall was identified. The dome of intramural segment was incised by knife and incision was prolonged until the real coronary artery orifice in the aorta was visualized. This real orifice matched in all cases the external vessel perpendicularly to the aortic wall. The intimal layer was reinforced with 7/0 polypropylene separate stiches. When the intramural course was below the plane of aortic valve commissures, the commissures were detached and then resuspended to the aortic wall. This maneuver was required only in one case in our experience.

If no intramural course but single coronary ostia was present two different techniques were used. In one case of AAOLCA a leftward repositioning of main pulmonary trunk was made at the beginning of our experience. Subsequently, in other three cases (2 AAOLCA and 1 AAORCA) the technique of neo-ostioplasty described by Vouhé and coll. [17] was used. When no intramural course but separate ostia were present the anomalous coronary artery was mobilized and re-implanted in the appropriate coronary sinus (2 AAOLCA and 1 AAORCA). Associated procedures were made in three patients (12%) patients and were the following: one patient with tricuspid insufficiency due to annular dilatation received a tricuspid valve repair by implantation of artificial ring. A second patient with associated large *ostiumsecundum* atrial septal defect underwent to concomitant closure of the ASD with autologous untreated pericardial patch. A third patients have a myocardial bridge associate to AAORCA. The myocardial bridge involved left interventricular artery and its extended for 20 mm in length and 4 mm in deepness. The patient underwent to myocardial muscular de-bridging by muscular unroofing.

Statistical analysis

Descriptive statistics was made and data were expressed as means with standard deviations or medians with ranges. Fisher exact test was used to compare preoperative clinical variables and surgical results between

the two main coronary subgroups (AAORCA vs AAORCA). Statistical analyses were performed using SPSS 24 (IBM Corp, Armonk, NY).

RESULTS

Patient's characteristics were summarized in table 1. Twenty-five patients underwent to surgical correction of AAOCA were included in the study. 17 (68%) were male. Median age at operation was 20 years (min 1 year – max 65 year). AAORCA were younger respect to patients with AAOLCA (median age 19 years vs 21, P = .11). AAORCA was diagnosed in 19 (76%) patients and AAOLCA was diagnosed in 6 (24%) patients. Two patients (8%) were already operated: one patient underwent previously coronary artery by-pass in a different hospital. He was subsequently, re-evaluated for chest pain persistence and AAOLCA was discovered after CT-scan. The other patient has been operated for ventricular septal defect closure.

Symptom of presentation and anatomic characteristics

Symptom of presentation were chest pain in 14/25 patients (56%). In all patients the chest pain was referred with the specific features of the cardiac chest pain (central in the chest and oppressive). Three patients (12%) were referred for ventricular arrhythmia, two patients (8%) were referred for non-specific cardio-vascular symptoms (dyspnea and dizziness). Six patients (24%) were asymptomatic and AAOCA was discovered during investigation for competitive sport. No episodes of sudden cardiac death were reported in our population. No statistical differences were found between AAORCA and AAOLCA for presence and type of symptoms of presentation (p =.09).

In hospital preoperative standard electrocardiographic investigation did not revealed ischemic alterations in all but ventricular arrhythmia was detected in three (12%). At routine preoperative echocardiographic investigation, two patients, both with AAORCA (8%) showed localized right basal hypokinesia.

Preoperative functional studies (treadmill test or nuclear perfusion scintigraphy) were made in 10/25 patients (40%). Five of them (20%), showed ischemic signs (two with AAORCA and three with AAOLCA, P = .17).

Computed tomography scan revealed the following anatomic characteristics: in the group of AAORCA, 15 (60%) patients had an inter-arterial and intra-mural course, while 4 (16%) had only inter-arterial course and right dominant coronary circulation. In the group AAOLCA, three (12%) patients had inter-arterial but no intramural course, two (8%) inter-arterial plus intra-mural course and one (4%) intra-mural course alone. In all but one cases the anomalous coronary artery originated from the opposite sinus (right coronary artery from left sinus and *vice-versa*) except for a case of AAOLCA in which the left coronary artery originated from non-coronary sinus and had intra-mural course. Median length of intramural segment was 8 mm (min 3 mm - max 11 mm).

Surgical results

Surgical unroofing was made in 18 patients (72%): 16 (64%) in AAORCA and 2 (8%), in AAOLCA. Coronary reimplantation was made in three patients (12%): two (8%) in AAORCA and one (4%) in AAOLCA. Coronary neo-ostioplasty was made in three patients (12%): two in AAOLCA and one (4%) in AAORCA. Pulmonary main trunk re-positioning was made in one case (4%) of AAOLCA with single ostium. No in hospital or long-term mortality was observed during the study period. No patients required re-operation for residual lesion, symptoms persistence or surgical related complications. Intensive care length of stay was less than 24 hours in all patients and mean hospital length of stay was 8,5 days. Hospital length of stay did not exceed 14 days.

Follow-up

Median follow-up was 5.3 years (min 9 months max 13 years). All patients received clinical examination, electrocardiogram and echocardiography. Computed tomography scan was scheduled after six months from surgery (12/25, 48% of cases). Exercise stress tests were made in 8/25 (32%).

All symptomatic patients reported the disappearing of the symptoms at follow-up and all athletes returned to

sport or physical activity. No basal ECG alteration were found in all. The echocardiographic studies showed normal ejection fraction and regional wall motion. No aortic insufficiency was observed in the patients required resuspension of the commissures. Both echocardiography and CT scan (when available) showed an improvement in the take-off angle (Figure 2). Exercise stress test were negative in all. No patients referred discomfort for surgical related pain (i.e. the sternotomy) or where scared about the restart of physical activity. All patients and their families were satisfied with the choice to have undergone to surgery.

DISCUSSION

AAOCA is the second common congenital heart malformation associated with sudden cardiac death [1,4]. Several anatomic variants have been described but AAORCA with inter-arterial and intra-mural course is the most frequently observed pattern. AAOLCA is less frequent but more associated with fatal events, especially in young athletes [1-4]. Other described coronary anomalies are the high origin of the coronary more than 1 cm above the sinu-tubular junction, the slit-like shape of coronary orifice or the common orifice for both left and right coronary arteries. These anomalies can be observed isolated or in association with the previously described [1].

The overall prevalence in the population of AAOCA is currently estimated between 0.1 and 1%. A recent improvement of cardiac screening in competitive athletes is presumably destined to increase their incidence in the next year [5]. Many groups, including our, have developed specific programs for the screening and management of AAOCA [11,16]. Risk stratification and indication to surgery, besides the anatomic evaluation, have encompassed different functional studies aiming to asses cardiac perfusion. Molossi and coll. showed interesting results with dobutamine stress cardiac magnetic resonance for assessment of myocardial perfusion and Angelini and coll. proposed IVUS (intra-vascular ultra sound) for anatomic and functional evaluation by dedicated guided catheters [18,19].

In the main time, guidelines and expert consensus have been developed and the debate on this topic was very prolific [8, 20,21]. Citing T.M. Sundt and M. Jacobs *"it is not the intended purpose of guidelines to take the place of clinical judgment and personalized care, but rather to provide a foundation that is a starting point for clinical decision making"* [22,23]. Based on this statement and looking to the current literature, it is our opinion, that at the moment indication to surgery are guided primarily by clinical and anatomical criteria [8,11,20,21,24]. In our experience a lot of symptomatic patients were observed among those with AAORCA (previously described as less symptomatic). Typical oppressive chest pain at rest and during efforts (sometimes not strenuous) was the most frequent symptom leading to further cardiologic investigation. Fifty-six percent of our patients referred typical oppressive chest pain while another 12% described palpitation. All of them showed ventricular arrhythmia at ECG. This results account for an overall frequency of cardiac symptoms of 68% in our series. 75% of these symptomatic patients had AAORCA (86% of these with inter-arterial and intra-mural course). These results, reinforced previous observation that severe cardiac symptoms may be present similar in AAORCA as in AAOLCA, despite functional test do not reveal cardiac ischemia [25]. In our population functional tests were negative in 50% of the patients who underwent to the studies. Probably, functional test may have the limit to not reproduce the dynamic condition of the heart's perfusion during sport's efforts or normal life (i.e. emotional stress or other factors that alter neuro-vegetative system), that continues to be the focus of the current research in AAOCA.

For these reasons, despite many efforts have been made to standardize the management of AAOCA, several groups continue to publish their own policy and results [12,12,13,24]. The lack of multi-centric prospective, randomized studies, ethnic and socio-economic differences, and the wide range of clinical presentation, probably hampers at the moment the possibility to define a standard management that can fits for all type of AAOCA.

Surgery have demonstrated to be safe in terms of mortality [14,15]. Freedom from reoperation as well was very low in all surgical published series. However, recurrence of symptoms is described up to 40% after surgical management [14,15,24]. Several techniques are used at the moment to approach AAOCA. Coronary unroofing seem to be the most simple and reproducible technique, and currently is the most adopted technique in the

United States [7]. However, this result may be due because intramural course is the more frequent anatomic pattern of AAOCA.

All patients with intra-mural course underwent to coronary unroofing at our institution regardless of type and length of AAOCA. In our experience with coronary unroofing we have focused our surgical technique in extending the opening of the roof until the "real" orifice of the coronary artery was detected to match perpendicularly the external take-off of the coronary from the aortic wall. Comparing pre-operative and post-operative imaging (echocardiography and CT scan) we have observed an improvement of the take-off angle (that frequently was associated with an enlargement of the internal diameter of the first segment of the vessel). Our results with coronary unroofing are comparable to those published by Mostefa-Kara and coll. that defined this technique as the "gold standard" for management of AAOCA [12]. However, despite results with coronary unroofing were excellent, there were different anatomic subtypes of AAOCA that in our opinion, are not suitable for this approach (i.e. absence of intramural course). Moreover, as described by Gharibeh and coll. the presence of intra-mural course is not an absolute recommendation for unroofing procedure. It is the overall neo-coronary shape that determines the improvement in coronary flow for these authors [26]. Our surgical policy aimed just to this point. Gaillard and coll. and Courand et coll. have showed optimal results in this way using neo-ostioplasty and coronary reimplantation as alternative technique for both left and right anomalous aortic origin of coronary artery, in children and young adults [13,25]. In our experience patients managed by neo-ostioplasty or coronary reimplantation showed overall same results to coronary unroofing. No patients developed cardiac related complications as ostial calcification sometimes described for neo-ostioplasty [13]. On the contrary, it was impressive, in our experience, the benefit of surgery on symptoms disappearing and this was the principal finding of our study. None of the symptomatic patients described symptoms recurrence postoperatively and at a median follow up of 5.3 years (min 9 months max 13 years). All young athletes returned to practice competitive sport.

The improvement of take-off angle probably reduces flow turbulence at coronary ostia. This mechanism improve perfusion and reduce ischemia. 4D flow MRI may be a useful method to validate this statement that at the moment is only a speculation, and may be the base for future investigation in this subgroup of patients. Previous study of fluid-dynamic has already demonstrated flow differences related to different surgical procedures [27]. Razavi and coll. have recently demonstrated how the improvement in the angle orifice is associated with a low coronary share stress and improved perfusion. This study is unfortunately limited only to coronary unroofing and do not account for different surgical techniques in order to compare them [28].

In conclusion, we believe that the disappearing of the symptoms and the return to sport activities may be considered as important outcome, besides mortality and freedom from reoperation, when we discuss surgical results and we want to offer surgery for AAOCA especially to young athletes.

Limits of our study are the retrospective nature of the research, the unnormal distribution of age of our patients and the lack of a complete long-term follow-up. Moreover, this represent a selected group of patients with the common features of an anatomic pattern described as at risk for SDC.

References:

1. Davis JA, Cecchin F, Jones TK, Portman MA. Major coronary artery anomalies in a pediatric population: incidence and clinical importance. J Am Coll Cardiol. 2001;37:593-7.
2. Poynter JA, Williams WG, McIntyre S, Brothers JA, Jacobs ML, ZiemerG,etal. Congenital Heart Surgeons Society AWG. Anomalous aortic origin of a coronary artery: a report from the Congenital Heart Surgeons Society registry.World J Pediatr Congenit Heart Surg. 2014;5:22-30.
3. Cheezum MK, Liberthson RR, Shah NR, Villines TC, O'Gara PT, Landzberg MJ, et al. Anomalous aortic origin of a coronary artery from the inappropriate sinus of Valsalva. J Am Coll Cardiol. 2017; 69:1592-608.
4. Angelini P. Sudden cardiac death: do we know what we are talking about? Circulation. 2002;105: E182
5. Mery CM, Lopez KN, Molossi S, Sexson-Tejtel SK, Krishanamurthy R, McKenzie D. Analysis to

define the optimal management of athletes with anomalous aortic origin of a coronary artery. J Thorac Cardiovasc Surg 2016; 152:1366-75
6. Brothers JA, Frommelt MA, Jaquiss RDB, Myerburg RJ, Fraser Jr CHD, Tweddel JS. Expert consensus guidelines: Anomalous aortic origin of a coronary artery J Thorac Cardiovasc Surg 2017; 153:1440-57
7. Jegatheeswaran A, Devlin PJ, Williams WG, Brothers JA, Jacobs ML, De Camply DW et al.

Outcomes after anomalous aortic origin of a coronary artery repair: A Congenital Heart Surgeons' Society Study. J Thorac Cardiovasc Surg 2020; 160:757-71

1. Jegatheeswaran A, Alsoufi B. Anomalous aortic origin of a coronary artery: 2020 year in review J Thorac Cardiovasc Surg 2021 Aug; 162 (2): 353-359
2. MoscaRS, PhoonCK. Anomalous aortic origin of a coronary artery is not always a surgical disease. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2016; 19:30-6.
3. Vouhe PR. Anomalous aortic origin of a coronary artery is always a surgical dis- ease. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2016; 19:25-9.
4. Mery CM, De Leon LE, Molossi S, Sexson-Tejtel K, Agrawal H, Krishnamurthy R et al. Outcomes of surgical intervention for anomalous aortic origin of a coronary artery: A large contemporary prospective cohort study J Thorac Cardiovasc Surg 2018;155:305-19
5. Mostefa-Kara M, Fournier E, Cohen S, Hascoet S, Van Aerschot I, Roussin R, El Zoghby J, Belli E. Anomalous aortic origin of a coronary arteries: is the unroofing procedure always appropriate? Eur J Cardiothorac Surg. 2021 Apr 13; 59(3): 705-710
6. Gaillard M., Pontailler M, Danial P, Moreau de Bellaing A, Gaudin R, du Puy-Montbrun L et al. Anomalous aortic origin of coronary arteries: an alternativevto the unroofing strategy. Eur J Cardiothorac Surg 2020; 58:975–82.
7. Jegatheeswaran A, Devlin PJ, McCrindle BW, Williams WG, Jacobs ML, Blackstone EH, et al. Features associated with myocardial ischemia in anomalous aortic origin of a coronary artery: a CHSS study. J Thorac Cardiovasc Surg. 2019; 158:822-34
8. Brothers JA, McBride MG, Seliem MA, Marino BS, Tomlinson RS, Pampaloni MH, et al. Evaluation of myocardial ischemia after surgical repair of anomalous aortic origin of a coronary artery in a series of pediatric patients. J Am Coll Cardiol. 2007;50:2078-82.
9. Bianco F, Colaneri M, Bucciarelli V, Surace FC, Iezzi FV, Primavera M et al. Echocardiographic screening for the anomalous aortic origin of coronary arteries. Open Heart 2021;8:e001495
10. Gaudin R, Raisky O, Vouhe´ PR. Anomalous aortic origin of coronary arteries: 'anatomical' surgical repair. Multimed Man Cardiothorac Surg 2014;2014: mmt022.
11. Molossi S, Agrawal H, Mery CM, Krishnamurthy R, Masand P. Outcomes in Anomalous Aortic Origin of a Coronary Artery Following a Prospective Standardized Approach Circ Cardiovasc Interv. 2020;13: e008445
12. Angelini P, Velasco JA, Ott D, Khoshnevis GR. Anomalous coronary artery arising from the opposite sinus: descriptive features and pathophysiologic mechanisms, as documented by intravascular ultrasonography. J Invasive Cardiol. 2003; 15:507-14.
13. Stout KK, Daniels CJ, Aboulhosn JA, Bozkurt B, Broberg CS, Colman JM et al. 2018 AHA/ACC guideline for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. Circulation. 2019;139: e698–e800
14. Baumgartner H, De Backer J, Babu-Narayan SV , Budts W, Chessa M, Diller GP et al 2020 ESC Guidelines for themanagement of adult congenital heart disease European Heart Journal (2020) 00, 1_83
15. Sundt TM. Guidelines or gospels? J Thorac Cardiovasc Surg. 2016; 151:1472-4.
16. ML Jacobs Anomalous aortic origin of a coronary artery: The gaps and the guidelines J Thorac Cardiovasc Surg 2017; 153:1462-5
17. Nees SN, Flyer JN, Chelliah A, Dayton JD, Tourchette L, Kalfa D et al. Patients with anomalous aortic origin of the coronary artery remain at risk after surgical repair J Thorac Cardiovasc Surg

2018;155:2554-64
18. Courand PY, Bozio A, Ninet J, Boussel L, Bakloul M, Galoin-Bertail C et al. Diagnosis and treatment of anomalous aortic origin of coronary artery: A twenty-year retrospective study of experience and decision-making in children and young adults International Journal of Cardiology 337 (2021) 54–61
19. Gharibeh L, Rahmouni K, Hong SJ, Crean AM, Grau JB. Surgical Techniques for the treatment of Anomalous Origin of Right Coronary artery from the left sinus: A comparative reviewe J Am Heart Assoc. 2021 Nov 16;10(22):e022377
20. de Leval MR, Migliavacca F, Guadagni G, Dubini G. Computational fluid dynamics in the evaluation ofhemodynamic performance of cavopulmonary connections after the Norwood procedure for hypoplastic left heart syndrome J Thorac Cardiovasc Surg 2003; 126:1040-7
21. Razavi A, Sachdeva S, Frommelt PC, La Disa Jr JF . Patient-Specific Numerical Analysis of Coronary Flow in Children with Intramural Anomalous Aortic Origin of Coronary Arteries. Semin Thorac Cardiovasc Surg 2021;33(1):155-167.

Table 1: Population characteristics

	All	AAORCA
N° of patients	25	19
Median age (years)	20	21
Male	17	15
Symptoms of presentation Chest Pain Arrhythmia Aspecific No symptoms	14(56%) 3 (12%) 2 (8%) 6 (24%)	10 2 1 4
Associate anomalies	2 (8%)	2
Interarterial course	24 (96%)	19
Intramural course	18 (72%)	15
Type of surgery Unroofing Re-implantation Neo.ostioplasty PT re-location	18 3 3 1	16 2 1 -
Mortality	0%	
Symptoms at Follow-up	None	None
Arrhythmia at Follow-up	None	None

AAORCA: anomalous aortic origin of right coronary artery; AAOLCA: anomalous aortic origin of left coronary artery

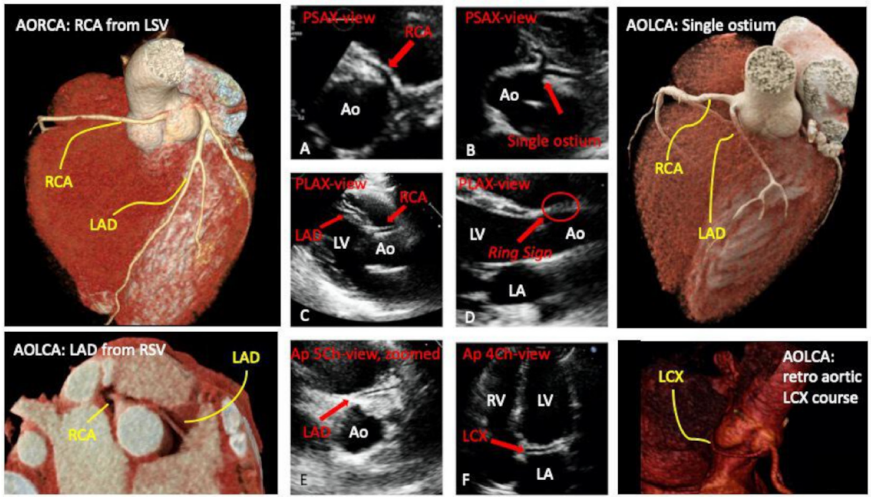

Figure 1: Two-dimensional echocardiographic 4-view approach for AAOCA detection: abnormal exami-

nations and their anatomic correlates. AO, aorta; AOLCA, anomalous origin of the left coronary artery; AORCA, anomalous origin of the right coronary artery; Ap-4Ch-view, apical four chambers view; Ap-5Ch-view, apical five chambers view; LA, left atrium; LAD, left anterior descending; LCX, left circumflex artery; LV, left ventricle; PLAX, parasternal long-axis view; PSAX, parasternal short-axis view; RCA, right coronary artery; RV, right ventricle; RVOT, right ventricular outflow tract (Reprint with permission from bianco and coll. Echocardiographic screening for the anomalous aortic origin of coronary arteries. Open Heart 2021;8:e001495)

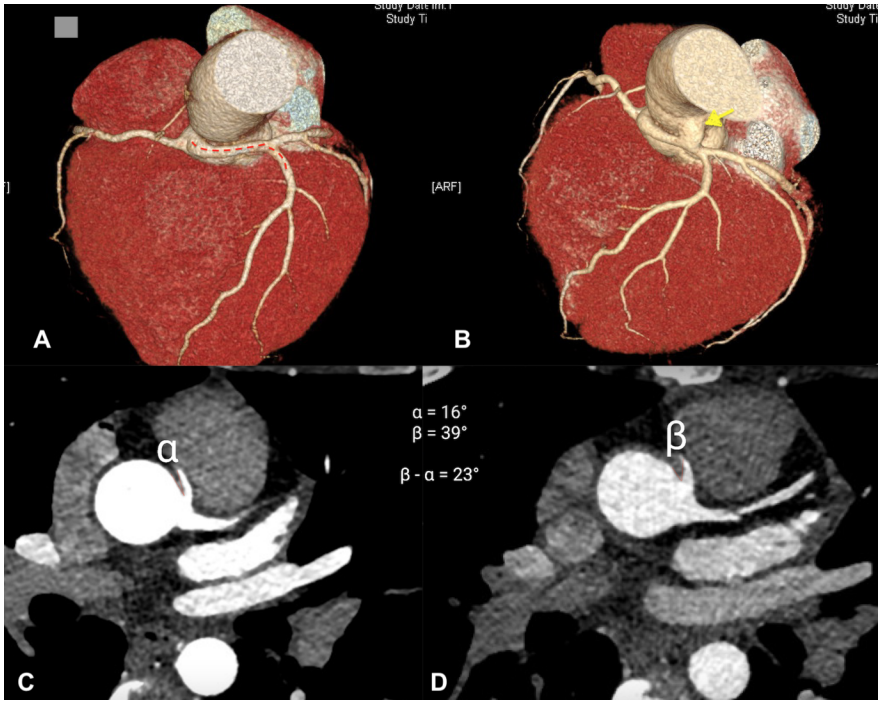

Figure 2: Computed tomography scan (CT) images showing the results of two different surgical techniques on arterial take-off angle. Upper panel (A-B): preoperative and postoperative CT three-dimensional reconstruction of AAOLCA (red dotted line) managed by Neo-ostioplasty - Vouhé technique (yellow arrow). Lower panel (B-C): CT axial view of preoperative (α) and postoperative (β) AAORCA managed by unroofing. It is possible to appreciate the more rectilinear origin of the arteries from the aorta.